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Case report

# A rare coexistence: Type 1 congenital intrahepatic portosystemic shunt in a patient with non-compaction cardiomyopathy☆,☆☆

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ABSTRACT

Non-compaction cardiomyopathy is described by the spongy appearance of myocardium and this condition frequently exists with other cardiac or extracardiac abnormalities. However, coexistence of non-compaction cardiomyopathy with congenital intrahepatic porto-systemic shunt was not reported previously. Here we presented a 22-year-old male patient with Type 1 congenital intrahepatic porto-systemic shunt and non-compaction cardiomyopathy.



## Case

A 22-year-old male patient was admitted to our clinic with fatigue and palpitation. He had no known personal or family history of cardiac disease. He was asthenic in appearance, a slight clubbing was observed on fingers and varicose dilatation of lower extremity veins was noted (Fig. 1, *Panel A*). Physical examination was otherwise normal. Electrocardiography showed left ventricular hypertrophy. Transthoracic echocardiography revealed normal left ventricular ejection fraction and increased apical trabeculations compatible with non-compaction cardiomyopathy and right ventricle was normal in appearance (Fig. 1, *Panel B*). Sizes of cardiac chambers were in normal limits and no significant valvular pathology was observed. However, on subcostal views, echocardiography showed an unexpected dilatation of inferior vena cava since right cardiac functions were normal. Evaluation with ultrasonography demonstrated an abnormal venous connection between portal vein and inferior vena cava (Fig. 1, *Panel C*). Blood test revealed normal ALT, AST, bilirubin and albumin levels and viral serology was negative for hepatitis viruses. Patient had no history of trauma, liver biopsy or surgery. Computed tomography and portal magnetic resonance venography confirmed type 1 congenital intrahepatic portosystemic shunt (Fig. 1, *Panel D*).

## Discussion

Non-compaction cardiomyopathy is described by spongy morphological appearance of the myocardium occurring particularly in the apical portion of the left ventricle. This condition is frequently associated with other cardiac and/or extra-cardiac (neurologic, visual and facial) abnormalities.[1] However coexistence of non-compaction cardiomyopathy with congenital intrahepatic porto-systemic shunt was not reported before. Congenital porto-systemic shunts are rare developmental abnormalities resulting in drainage of porto-mesenteric blood into systemic circulation partially or totally by passing the liver. Development of portal venous system occurs 4th to 10th week of embryogenesis and congenital anomalies of portal venous system are derived from abnormal coalescence of the vitello-umbilical venous plexus. Park et al. described 4 types of intrahepatic shunts. According to this classification; in Type 1 shunt one large connection between portal vein and inferior vena cava, in Type 2 shunt one or more connection between peripheral branches of portal vein and hepatic vein, in Type 3 shunt an aneurysmal connection between peripheral portal vein and hepatic vein, in Type 4 shunt multiple connections between portal vein and hepatic vein are

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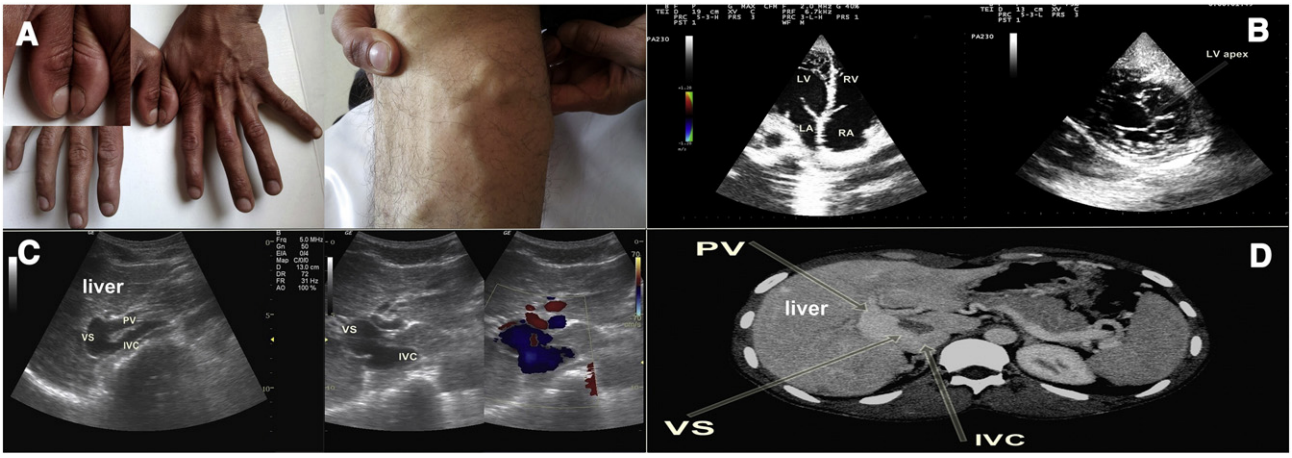


**Fig. 1.** *Panel A*. Slight clubbing of fingers and varicose veins on lower extremity. *Panel B*. Apical four chamber and parasternal short axis view showing hypertrabeculation of left ventricle compatible with non-compaction cardiomyopathy. LA; left atrium, LV; left ventricle, RA; right atrium, RV; right ventricle. *Panel C*. Ultrasonography showing an abnormal venous connection between portal vein and inferior vena cava. *Panel D*. Computed tomography portal venography showing intrahepatic portosystemic shunt. IVC; inferior vena cava, PV; portal vein, VS; venous shunt.

described.[2] Although the detection of portosystemic shunt in this patient might be incidental finding, possibility of association of non-compaction cardiomyopathy with other vascular abnormalities would not be underestimated. On the other hand, several growth factors (i.e. vascular endothelial growth factor), intracellular signaling pathways and, cell adhesion molecules (i.e. vascular cell adhesion molecule-1) play critical role in both left ventricular non-compaction and portosystemic shunt development.[3,4] As known, heart muscle is non-compacted between 4th and 18th week of development and any environmental or genetic impact during this period which may affect the common processes of differentiation in myocardium and vitello-umbilical venous plexus may result in both non-compaction cardiomyopathy and congenital porto-systemic shunts. Noticing the subcostal views in cases with non-compaction cardiomyopathy may provide the initial clue for such rare coexistences.

## Conflict of interest

None declared.

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